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CONGENITAL MINI-FOCUS ISSUE

ADVANCED

CASE REPORT: CLINICAL CASE

HeartMate III as a Bridge to Transplantation in an Adolescent With Failed Fontan Circulation

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ABSTRACT

HeartMate III is an emerging, small-sized centrifugal ventricular assist device. Its lower pump thrombosis and stroke rates make it favorable for use in pediatrics. We report the use of HeartMate III as a bridge to transplantation in an adolescent with failed Fontan circulation. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2019;1:512–5)
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HISTORY OF PRESENTATION

A 14-year-old male with hypoplastic left heart syndrome was admitted with shortness of breath and abdominal pain. He was found to have heart failure with desaturation, ascites, hepatomegaly with elevated liver enzymes, and renal dysfunction. His troponin and N-terminal pro-brain natriuretic peptide levels were elevated. Echocardiography showed severely depressed systemic ventricular function. Cardiac catheterization showed thrombus in the native aortic root with compromised flow into both coronaries. Thrombolytic therapy was initiated with near resolution of thrombus. He continued to be inotrope-dependent and experienced a bradycardic arrest that required cardiopulmonary resuscitation. His ventricular function remained severely depressed; he was transferred to our institution on high-flow nasal cannula, milrinone, and dopamine for heart transplant (HTx) evaluation.

LEARNING OBJECTIVES

- VAD support is a feasible option for patients with failing SV physiology as a BTT.
- HeartMate III is a low-profile inflow cannula pump that has higher survival and lower pump thrombosis and/or stroke rates compared with other VADs, which makes it favorable for use in pediatrics.

MEDICAL HISTORY

The patient initially underwent a Norwood/Sano stage I procedure and subsequently extracardiac nonfenestrated total cavopulmonary connections. He was lost to follow-up before presenting with progressive oxygen desaturation and decline in ventricular function.

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Informed consent was obtained for this case.

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INVESTIGATIONS

The patient was initially placed on a veno-arterial extracorporeal membrane oxygenator (VA-ECMO) before proceeding with ventricular assist device (VAD) placement due to worsening heart failure on dual inotropic therapy with progressive hemodynamic instability and end-organ dysfunction (peak alanine aminotransferase/aspartate aminotransferase 150/92 U/l, respectively; creatinine 1.93 mg/dl). The patient was listed as status 1A on the HTx waitlist, but meanwhile underwent chest computed tomography angiography to facilitate 3- and 4-dimensional reconstructions to determine ideal placement of the VAD (**Figure 1**). On day 6 of VA-ECMO support, patient underwent re-sternotomy, and the VA-ECMO arterial cannula was exchanged with direct ascending aorta cannulation because of bleeding from the leg. The chest was left open with a plan for elective VAD placement in subsequent days.

MANAGEMENT

On day 8 of VA-ECMO, the patient was brought to the operating room, and cardiopulmonary bypass was established. On beating heart, under transesophageal echocardiography (TEE) guidance, part of the diaphragmatic surface of the right ventricle was cored out, and the cavity was inspected. Part of the tricuspid valve leaflets and subvalvar apparatus were removed due to proximity to the VAD inflow cannula. The inflow cannula of the VAD was inserted and secured with multiple interrupted pledgeted 2/0 ethibond sutures. The outflow graft was anastomosed to the side of the neo-aorta with 4/0 prolene sutures. De-airing was achieved. The pump speed was slowly increased to 5,200 rpm and stabilized at this level. The patient was weaned off cardiopulmonary bypass without inotropic support. Flow was stabilized around 3 l/min with a pulsatility index of 3.6. Postoperative chest x-ray and TEE showed adequate VAD placement (**Figures 2 and 3**).

FOLLOW-UP

The VAD parameters remained stable (**Figure 4**). Over 2 weeks, the VAD speeds were slowly increased to achieve flows of 4 l/min and to avoid low-flow events due to an excessive suctioning effect (**Figure 4**). Bivalirudin was used as initial VAD systemic anticoagulation because of its rapid enzymatic reversibility and titratability in this patient who had earlier hemostasis issues and evolving hepatic and renal function after implantation. Low-dose bivalirudin (0.02 mg/kg/h) was initiated and titrated up to maximum infusion of 0.3 mg/kg/h to a target activated partial thromboplastin time of 55 to 70 s and an international normalized ratio of 2.0 to 3.0. After enteral feedings were established, he was transitioned to warfarin that targeted factor X levels of 20% to 40% (corresponding to an international normalized ratio of 2.0 to 3.0) while simultaneously titrating his bivalirudin down to maintain an activated partial thromboplastin time at 60 s. Aspirin was also administered to maintain a therapeutic Verify Now (Accumetrics, Inc., San Diego, California) of <550. Ultimately, the patient did well and was discharged on post-VAD implantation day 34 with VAD flows of 4.8 l/min. The patient did well at home in the interval between VAD implant and HTx. He underwent successful HTx on day 43 post-VAD implantation.

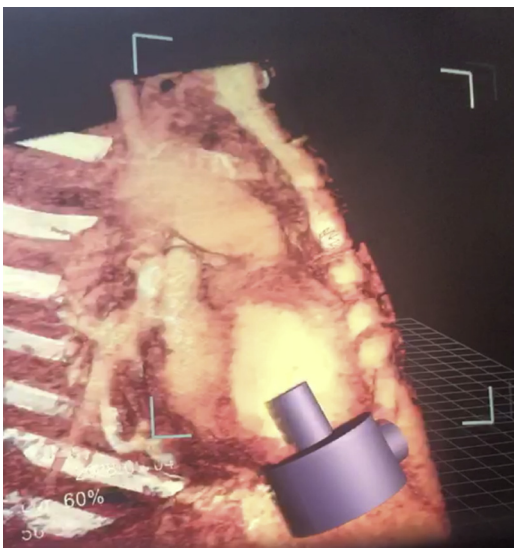
DISCUSSION

The Fontan procedure is the final palliation for patients with single ventricle (SV) physiology. However, there are well-known, long-term complications associated with Fontan physiology, such as restricted exercise capacity, systemic and/or hepatic venous hypertension, portal hypertension, pulmonary arteriovenous malformations, veno-venous shunts,

ABBREVIATIONS AND ACRONYMS

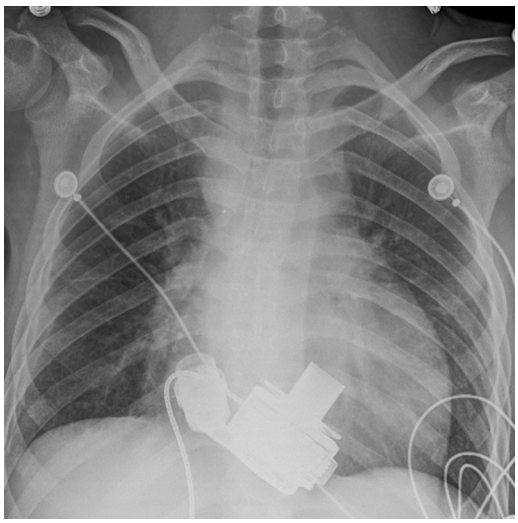
BTT = bridge to transplant
DT = destination therapy
HTx = heart transplant
SV = single ventricle
TEE = transesophageal echocardiography
VAD = ventricular assist device
VA-ECMO = veno-arterial extracorporeal membrane oxygenator

FIGURE 1 Virtual 3D VAD



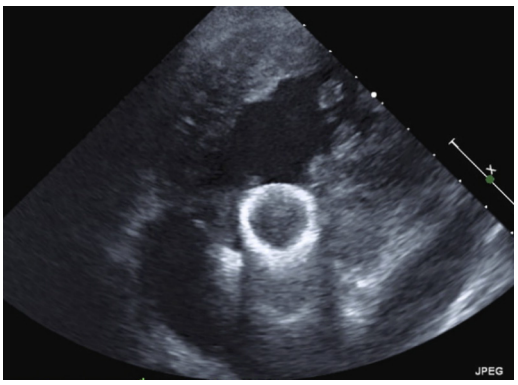
Three-dimensional (3D) image using the EchoPixel system (Boston Scientific, Marlborough, Massachusetts) with virtual superimposition of Heartmate III. VAD = ventricular assist device.

FIGURE 2 Post-Implantation HeartMate III X-Ray



Chest roentgenogram demonstrating adequate HeartMate III device position into the right ventricle.

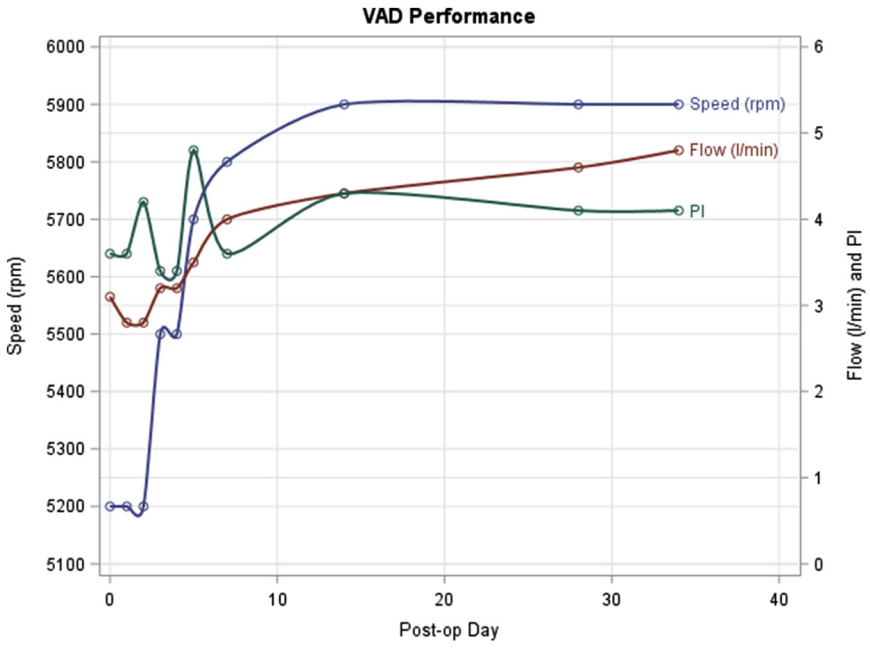
FIGURE 3 TEE Image



Inflow cannula position and decompression of the systemic right ventricle. TEE = transesophageal echocardiography.

lymphatic dysfunction, and systemic ventricular failure (1). Waiting time for transplantation has been a major limitation, particularly in patients with congenital heart disease who may be highly sensitized. VADs have emerged to bridge this gap and to improve clinical status of patients before HTx. VAD use has been reported after all stages of SV palliation; Vander Pluym et al. (2) presented extremely high mortality (nearly 40%) in their cohort. To improve the quality of care for patients with SVs with VAD support, the MFICS (Mechanical Support as Failure Intervention in Patients with

FIGURE 4 VAD Performance



Speed in rotations per minute (rpm), flow in liters per minute (l/min), and pulsatility index (PI) in LPM. Abbreviation as in Figure 1.

Cavopulmonary Shunts) registry was established (3). Lorts et al. (4) introduced the first case of HeartMate III (Abbott, Chicago, Illinois) implantation in an adult with a failing Fontan.

VA-ECMO is a feasible short-term option; however, complications can develop within weeks, which makes it an unsuitable device for bridge to transplant (BTT) (5,6). In addition, VA-ECMO has high morbidity and mortality post-HTx (7). For longer term support, a different set of devices has emerged. These include Berlin Heart EXCOR (Berlin Heart GmbH, Berlin, Germany), HeartMate II (Abbott-Thoratec, Chicago, Illinois), HeartWare HVAD (HeartWare International, Framingham, Massachusetts) and HeartMate III (Abbott).

HeartMate III is used as BTT or destination therapy (DT) in patients with end-stage heart failure (8). However, in the pediatric population, DT is not a primary option, and most devices are used to BTT. It has been associated with lower incidence of pump thrombosis and stroke rates compared with both the HeartWare HVAD and HeartMate II (8,9). United States approval for HeartMate III was supported by the Momentum 3 trial. Patients with the HeartMate III left ventricular assist device had an unprecedented survival rate (77.9%), very low pump thrombosis (1%), and the lowest stroke rate (10%) at 2 years (8). Despite being off-label use for pediatrics, this device was chosen as BTT therapy for this case because of the pump size, the lower complication rates, the body surface area of the patient (1.5 m^2), and the need for mid-term support because of the patient's blood type and human leucocyte antigen sensitization. In addition, because the patient had bleeding complications on ECMO, the authors elected to use HeartMate III because it could be used with lower anticoagulation goals than the HeartWare device.

HeartMate III is a centrifugal flow pump with a low profile inflow cannula, which may make this device favorable in SV anatomy. In this patient, the authors removed additional right ventricle and tricuspid valve tissue to clear the area for the inflow cannula. The authors used lower pump speeds than those typically reported coming out of the operating room to allow gentle flow adaptation of the Fontan pulmonary passive circulation in the early post-implantation stages. Fontan circulation seemed to be well supported with this device, with adequate end-organ perfusion with resolution of renal and/or hepatic dysfunction and a decrease in central venous pressures. Ventricular diastolic volume decreased from 147 ml pre-VA-ECMO and pre-VAD implantation to 105 ml post-VAD implantation.

CONCLUSIONS

The authors believe Heartmate III can be considered for BTT therapy, with lower complication rates for pediatric patients with failing Fontan physiology. The initial low flows on the VAD are essential with steady increases over time and are better tolerated in patients with SV physiology because the Fontan circulation needs to adapt to the increase of cardiac output over time.

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